



**CASE REPORT**

**CHOROIDAL MELANOMA IN PATIENT WITH VITILIGO: CASE REPORT**

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**ABSTRACT**

Choroidal melanoma is the most common primary intraocular malignancy in adults , arising from melanocytes within the uveal tract. Vitiligo is an autoimmune skin disorder characterized by depigmentation due to melanocyte destruction . While vitiligo has been associated with a decreased incidence of cutaneous melanoma , reports of uveal melanoma in patients with vitiligo remain rare . This article presents a case of choroidal melanoma in a patient with a known history of more than 20 years vitiligo, explores the possible immunological implications of this association and shows the ophthalmological management of such a rare case.

**Keywords:** Choroidal melanoma, intraocular malignancy vitiligo, ophthalmological management

**INTRODUCTION**

Choroidal melanoma is the most common malignant neoplasm occurring intraocularly, and it is the second most common neoplasm after cutaneous melanoma, arising from the melanocytes of the uveal tract<sup>1,4</sup>. The causes of its occurrence are still unknown, but oxidative damage to tissues and genetic factors have a significant impact<sup>5</sup>. Vitiligo affects approximately 0.5-2 % of the global population and is marked by selective loss of epidermal melanocytes . Autoimmune mechanisms are strongly implicated in its pathogenesis<sup>6</sup>. Interestingly , some studies suggest that vitiligo may confer protection against melanoma due to heightened immune surveillance against melanocyte antigens<sup>2,3</sup>. However, such rare cases of uveal melanoma in patients with vitiligo challenge this hypothesis.

**CASE PRESENTATION**

A 45-year-old woman presented to the with a 20-year history of generalized vitiligo presented to the Department of Oculoplastic and Neuro-

of phthalmological center after S.V. Malayan with progressive visual loss of right eye during last month(fig1a,b).



**Figure 1.(a)** Depigmentation of skin, hair roof, polios

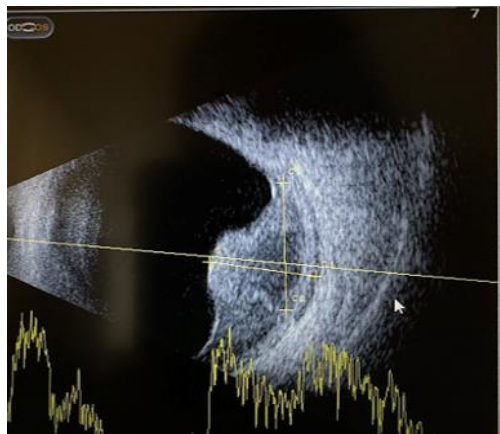


**Figure 1.** (b) Depigmentation of skin, hair roof, poliosis

The external examination showed a total absence of skin and hair roof pigment, particularly in the periorbital areas, poliosis. Patient also reported that she had vitiligo for over 20 years, but she was not examined by dermatologist and that was suspected diagnosis. On general ophthalmological examination visual acuity in the right eye was eccentric 0.01: 20/2000, in the left eye 1.0: 20/20. The intraocular pressure in the right eye was 12 mm, and in the left eye: 17 mm ( by pneumotonometry ). Ophthalmoscopy revealed a pigmented choroidal mass suggestive of melanoma in the right eye. So patient was underwent ocular ultrasound of right eye Fundus photography of right eye showed a dome-shaped, pigmented intraocular lesion.



**Figure 2.** Fundus photography of left eye showed general palor of fundu



**Figure 3.** B-scan of right eye

Ocular ultrasound answer was destruction of the vitreous body, 9.13 mm prominence, which has a base of 11.21\*11.84 mm: + tissue, secondary retinal detachment and subretinal hemorrhage. The patient underwent to CT-scan and MRI. The MRI also revealed a tumor in the right eyeball measuring 8\*8 mm: melanoma.



**Figure 4.** MRI Scan of head and orbit

Patient was underwent to CT Scan of abdominal and pelvic organs by gynecologist due to complaints of uterine bleeding. The CT-csan showed a massive myoma of the uterus measuring 14.50\*12.1\*12.7 mm, and no other pathological changes were found in the parenchymatous organs. Based on all the results of the clinical and radiological examinations, a diagnosis of choroidal melanoma in the right eye was made. Due to the sizes of tumor was planned enucleation with implantation of right eye under general anesthesia. Preoperative examinations revealed iron deficiency anemia - Hemoglobin degree 62.0 g/l (120.0 - 150.0 g/l) Ferritin degree 2,56 ng/ml (20.0 - 200.0 ng/ml), which was a direct contraindication for the surgery, and the patient was underwent to the hematologist for treatment. Two months later, during a follow-up visit, the visual acuity of the right eye had become perception of light with correct projection. Repeated ultrasound showed a significant increase in the sizes of the melanoma, measuring 10.30 mm in prominence and 14.65\*12.50 mm at the base.

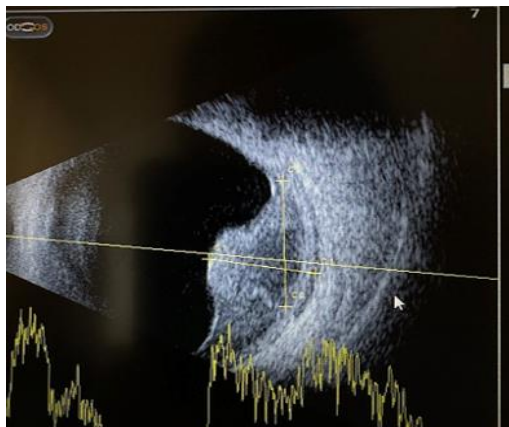


Figure 5. 1st B-scan of right eye

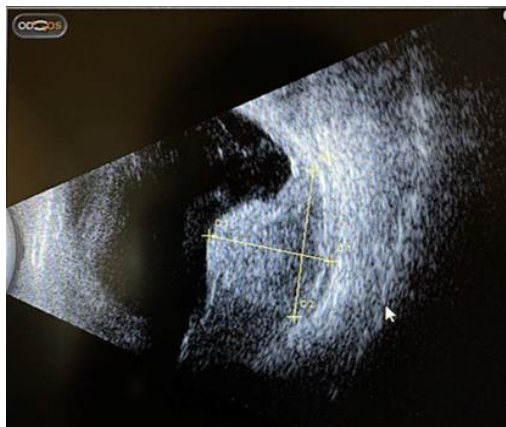


Figure 6. 2nd B-scan of right eye

During this time, a hysterectomy was performed due to huge myoma of uterus, the hemoglobin level is now regulated 123 g/l (120.0 -150.0 g/l) , and there were no therapeutic and anesthesiologic contraindications for surgery. Therefore, considering the location of the melanoma, the significant increase in size, progressive visual lost, the absence of object vision, enucleation was performed with non-integrated silicone spherical implant and conformer implantation

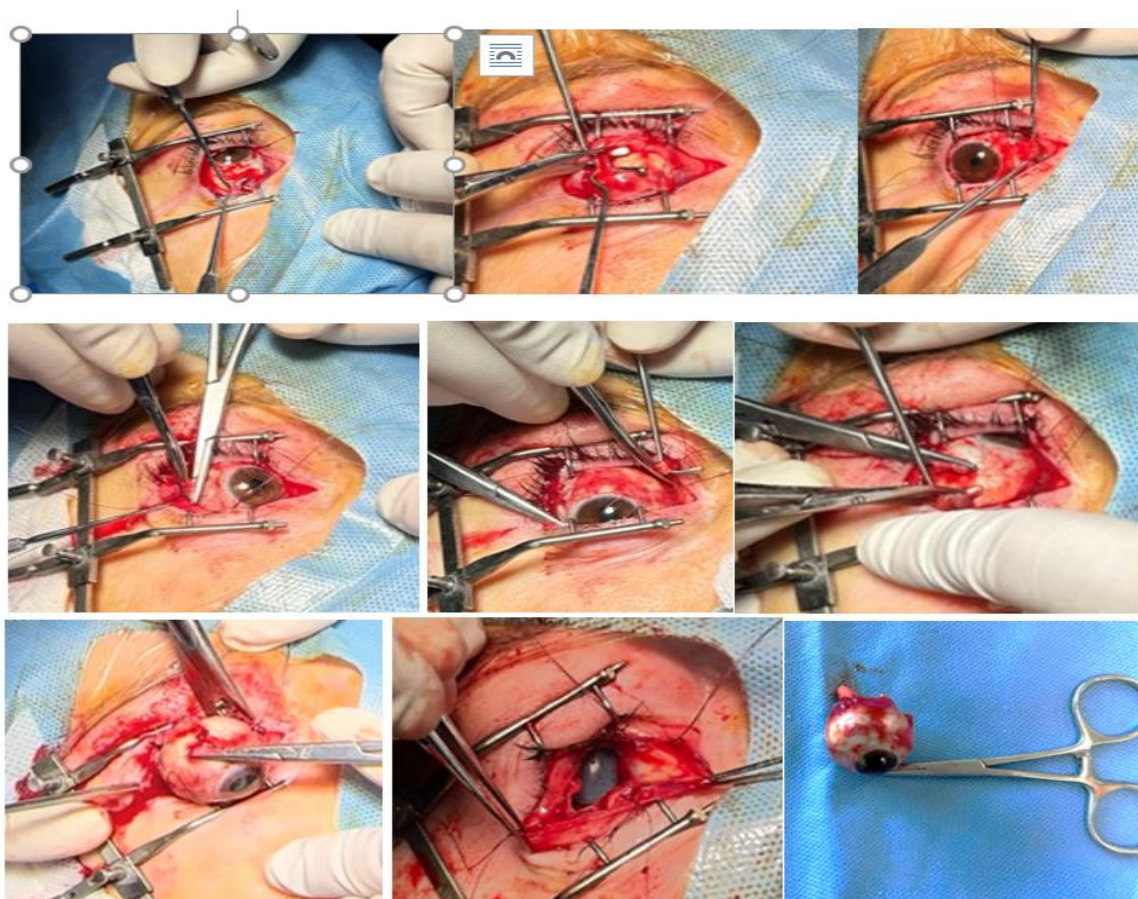


Figure 7. The surgery: Enucleation of right eye: step by step: Surgeon Armine Gharakeshishyan MD Head of the Department of Oculoplastic and Neuro-Ophthalmology, Ophthalmological center after S.V. Malayan, Yerevan, Armenia



Figure 8. Post-op 7th day

Figure 9. Post-op 14th day

On the 7th post-operative day we already had the answer of the histological examination invasive uveal melanoma of right eye the pigmented form, surgical radicality was completed.

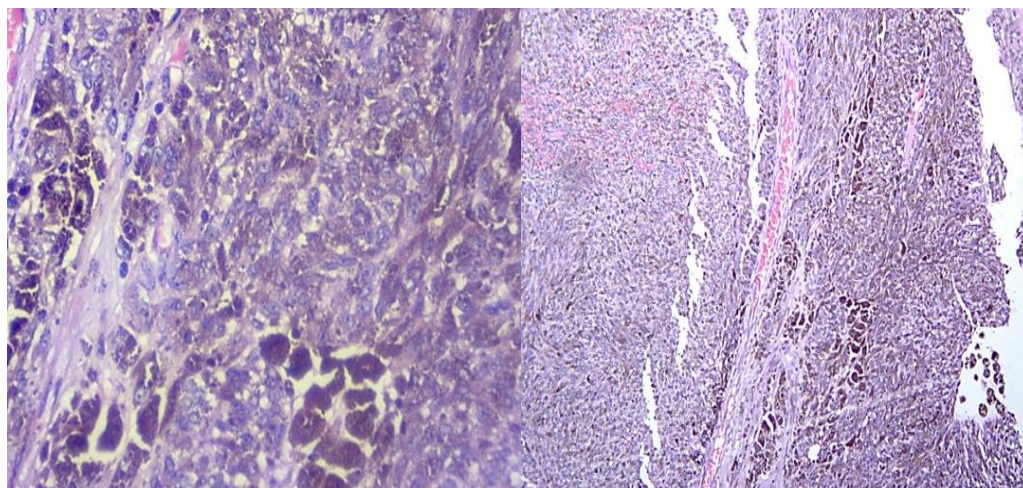


Figure 10. Histological slides: Histological section stained with H&E showed pigmented form of uveal melanoma: Histologist D. Mamunts, Davidyants laboratories.

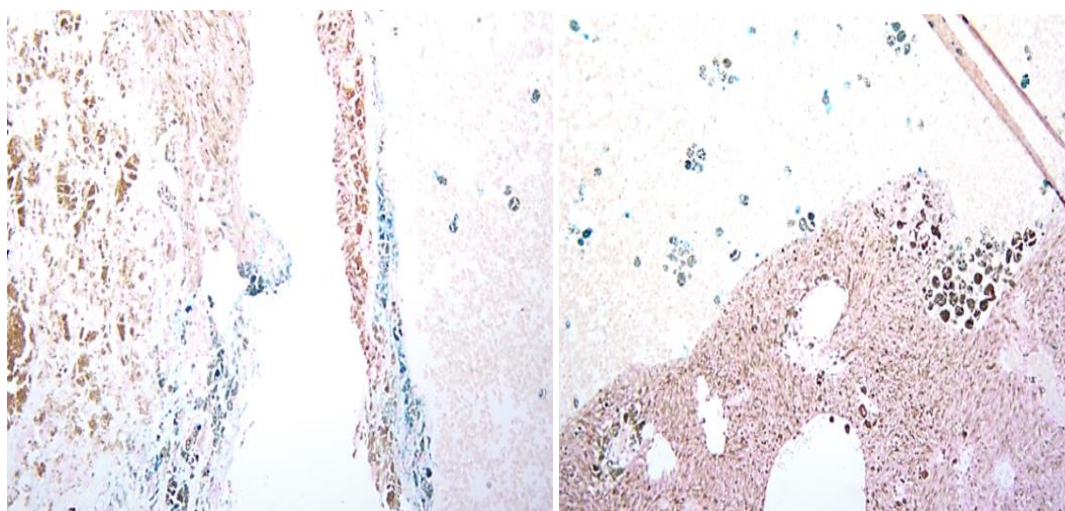
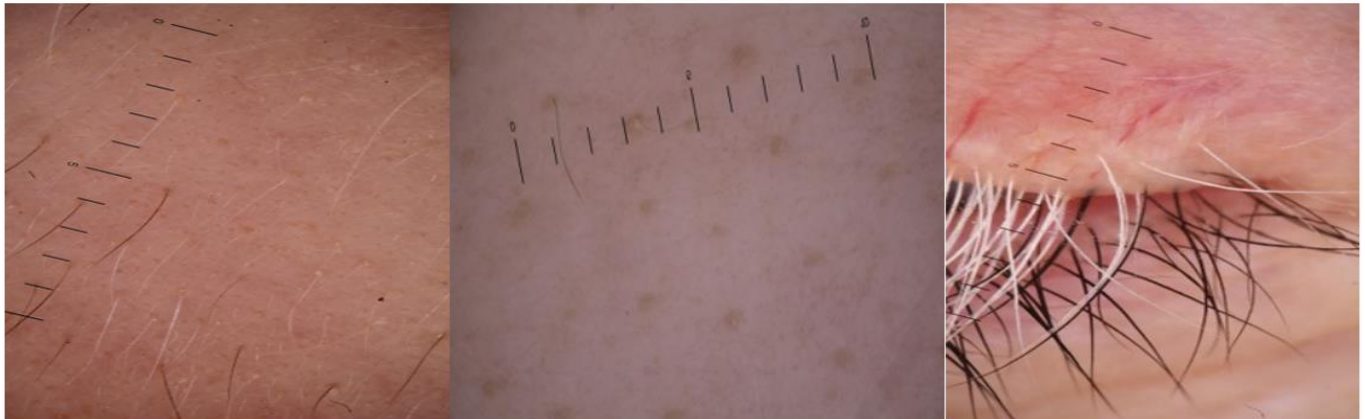


Figure 11. Histological slides: Histological evidence of melanoma : Histological section stained with Perls' Prussian blue demonstrated melanocytes in brown color, hematocytes in blue color. Histologist D. Mamunts, Davidyants laboratories RA

Since the patient didn't provide a clear basis for the diagnosis of vitiligo, and there were almost no documented similar cases of patients with vitiligo in literature, there were cases of melanoma development in patients with albinism. Therefore, it was decided to refer the patient to dermatologist's consultation and was performed dermatoscopy for clinically and scientifically confirm the diagnosis of vitiligo. According to the clear dermatoscopic criterias, the diagnosis of total vitiligo was confirmed.



**Figure 12.** Dermatoscopy slides: Dermatoscopy criteria's: Development of color grid pattern, lack of color grid pattern, white uneven spotted bands, perifollicular hyperpigmentation, branched linear vascular structures, leukotrichia. Dermatologist Shushanik Karapetyan, National center of dermatology. The patient was under the oncologist's supervision and there was no pathology in other organ systems. In 6st postoperative week the patient was send for final cosmetic prosthetics



**Figure 13.** The patient in postoperative period, final cosmetic prosthetics

## DISCUSSION

Choroidal melanoma is the most common malignant neoplasm occurring intraocularly, and it is the second most common neoplasm after cutaneous melanoma, arising from the melanocytes of the uveal tract<sup>4</sup>. 85 % of ocular melanoma originate in the uveal tract, 10% originate in melanocytes of epithelium of conjunctival membrane. The mortality is 45 %<sup>5</sup>.

Pathophysiology is still unclear, the mane cause is oxidative damage of pigmented tissues, genetic factors. Risk factors are ultraviolet exposure,<sup>5</sup> light

skin, hair, light eye iris color, iris nevus, nevus of Ota, abnormal cutaneous nevus, dysplastic nevus, heterochromia, freckles, outdoor activities, tanning.<sup>5</sup> The signs and symptoms are non-specific, depend on tumor location and sizes, usually they are blurry vision, flashing lights, visual field defects, metamorphopsia, some pain or can go asymptomatic<sup>5</sup>. Classification by sizes : small 4.0-8.0 mm diameter / 1.0-2.4 mm height, medium 6-16 mm. diameter / 2.5-10.0 mm height, large more 16 mm diameter / > 10.0 mm height ; by cell type:epithelioid,spindle, mixed; by staging system:the

tumor, node,metastasis<sup>5</sup>. The diagnostics is fundus examination, fundus photography, A-scan, B-scan ultrasound, fluorescein angiography, optical coherence tomography, magnetic resonance imaging, computed tomography, positron emission tomography<sup>5</sup>.Management is surgical or non-surgical. Surgical methods are enucleation or exenteration, local surgical resection, transscleral resection<sup>4,5</sup>. Non-surgical eye salvage treatment is radiation, brachytherapy, plaque radiation, particle beam radiotherapy , transpupillary thermotherapy, laser photocoagulation, systemic chemotherapy. Surveillance : 5 – year relative survival rate is 70-80 % , 20 -30 % of UM patients die within 5 years,45% die within 15 years.<sup>5</sup> 50 % of UM will have metastases by hematogenous spread to liver 90%, lungs 24%, bones 16%. Vitiligo affects approximately 0.5-2 % of the global population and is marked by selective loss of epidermal melanocytes<sup>6</sup>.

Autoimmune mechanisms are strongly implicated in its pathogenesis<sup>2,3</sup>. Interestingly, some studies suggest that vitiligo may confer protection against melanoma due to heightened immune surveillance against melanocyte antigens<sup>7</sup>.However, such rare cases of uveal melanoma in patients with vitiligo challenge this hypothesis. And this case was very challenging for both ophthalmologists, oncologists and histologists, as we had a patient with vitiligo, which means absence of melanocytes, but she got the pigmented form of choroid melanoma which origins from melanocytes. So in our practice we had this very rare case, and it deserves discussion and a place in the literature.

## CONCLUSION

Although rare, choroidal melanoma can occur in patient with vitiligo. This case underscores the need for ophthalmic evaluation in vitiligo patients with visual symptoms and encourages further investigation into the immunological interplay between autoimmunity and melanocytic malignancy. Considering the risk factors, preventive measures can be taken. It usually is diagnosed unexpectedly. Choroidal melanoma in a patient with vitiligo is a very rare case, there was only one case described in the literature involving an Asian patient; in all other articles, vitiligo was developed in patients with melanoma on the background of immunotherapy.

## DECLARATION

### Ethics approval and consent to participate

The study received ethical approval from the institutional the Ethics Committee

### Consent for publication

Not applicable.

### Competing interests

The authors declare no conflict of interest.

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